

Rare ocular manifestations in an 11-year-old girl with incomplete Kawasaki disease

A case report

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Abstract

Introduction: Kawasaki disease is a necrotizing vasculitis featuring fever, erythema, conjunctivitis, and lymphadenopathy. Ocular manifestations in Kawasaki disease are commonly limited to anterior segment, posterior segment lesions are rarely reported.

Case presentation: We report a unique case of ocular manifestations in an 11-year-old girl with incomplete Kawasaki disease. An 11-year-old Asian girl presented with severe enophthalmos, retinitis, retinal detachment, and choroidal detachment secondary to an unexplained fever for 10 days.

Conclusion: To the best of our knowledge, this is the first documented case of incomplete Kawasaki disease with severe posterior segment lesions. The local use of dexamethasone in the eye was effective in our patient. Surgical intervention might not be necessary even though the initial symptoms could be devastating. The eye should be monitoring the eye routinely in patients with Kawasaki disease.

Abbreviations: IOP = intraocular pressure, IVIG = intravenous immunoglobulin.

Keywords: Kawasaki disease, ocular manifestations, retinal detachment, retinal vasculitis

1. Introduction

Kawasaki disease is a necrotizing vasculitis featuring fever, erythema, conjunctivitis, and lymphadenopathy.^[1] Ocular manifestations in Kawasaki disease are commonly limited to anterior segment characterized by bilateral bulbar conjunctivitis without exudate, superficial punctate keratitis, uveitis, or vitreous opacities.^[2]

We found 6 literatures^[3–7] describing posterior segment lesions in Kawasaki disease. But none of them progressed to retinal detachment. Only one literature^[7] described a patient lost vision in 1 eye due to retinal and vitreous exudation. We report a unique case of incomplete Kawasaki disease (atypical) with severe enophthalmos, cataracts, exudative retinal detachment, and retinal vasculitis.

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YG and YZ have contributed equally to this work.

Written informed consent was obtained from the parent for publication of this case report.

This case report is not a clinical trial, so ethical approval was not necessary.

All the authors report no conflict of interests.

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2. Report of a case

An 11-year-old Asian girl presented with a 10-day history of fever (up to 40.5°C), generalized rash, erythema of oral mucosa, cervical lymphadenopathy, bilateral bulbar conjunctivitis without exudate, and cervical lymphadenopathy. Laboratory results showed elevated white blood cell count with neutrophil predominance C-reaction protein, erythrocyte sedimentation rate, platelet count, serum liver enzymes, and decreased albumin. Patient did not have strawberry tongue, erythema, and edema of the hands and feet. Electrocardiogram showed no abnormal findings. Incomplete (atypical) Kawasaki disease was suspected (Table 1).^[1,8–11]

Patient was immediately treated with intravenous immunoglobulin (IVIG) for 10 days in a community hospital. Unfortunately, she responded poorly to the treatment, the fever persisted despite the infusion of immunoglobulin. Ten days later, she was transferred to our university hospital. We added high-dose corticosteroids to her treatment regimen.

Table 1

Diagnosis of atypical Kawasaki disease^[1,8]

A diagnosis of atypical Kawasaki disease can be made in the presence of both of the major symptoms and a positive echocardiogram^[9] or ≥3 minor symptoms.

Major symptoms:

1. Children with fever >5 days and 2 or 3 compatible clinical criteria or infants with unexplained fever for >7 days^[10]
2. CRP >3.0 mg/dL and/or ESR >40 mm/h^[11]

Minor symptoms:

1. Anemia for age
2. Platelet count of >450,000 after the 7th day of fever
3. Albumin <3.0 g/dL
4. Elevated ALT level
5. WBC count of 15,000/mm³
6. Urine >10 WBC/hpf

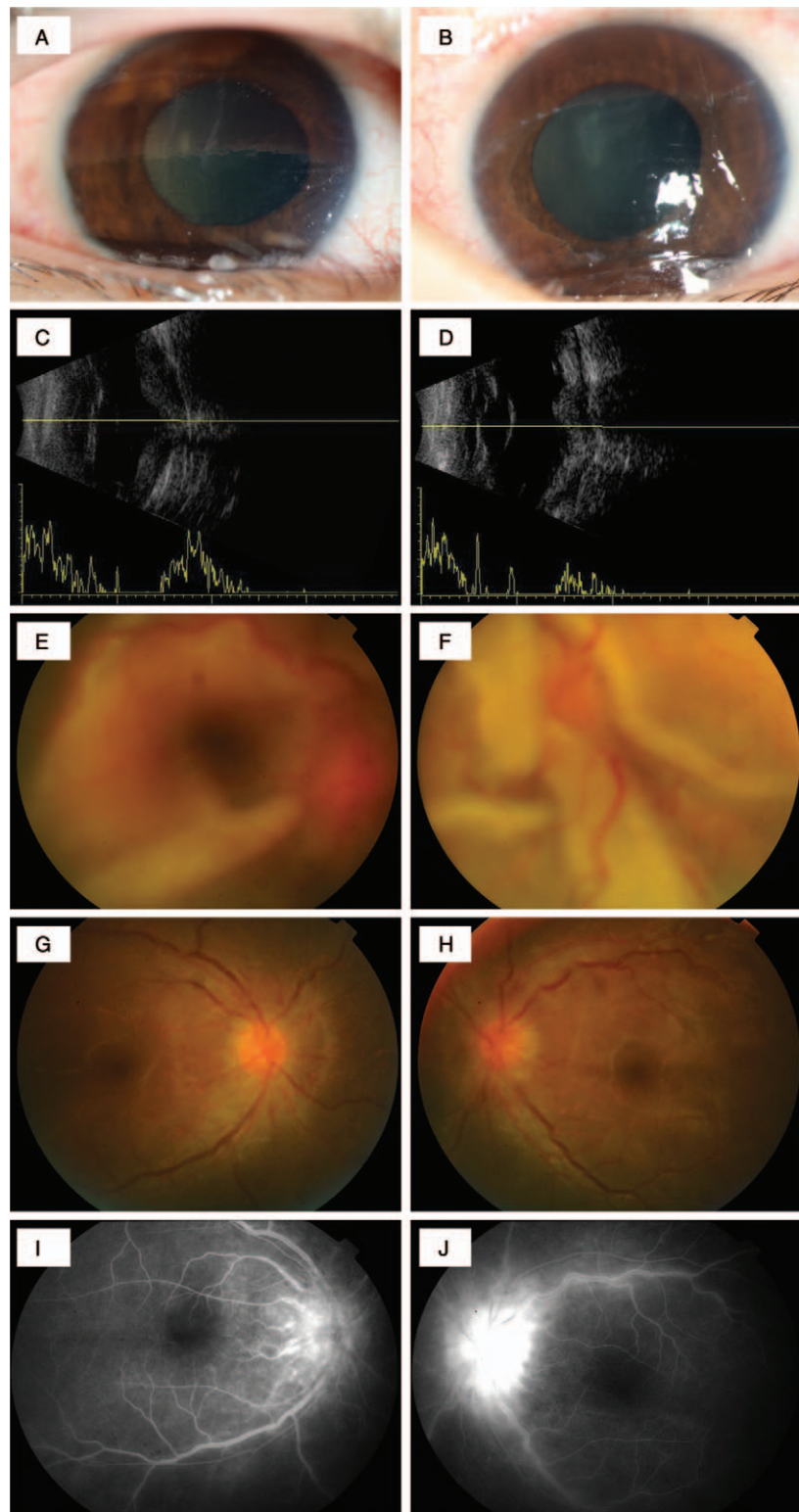


Figure 1. A, Photograph of the right eye, and B, photograph of the left eye showed severe deformation, conjunctival edema, and injection without exudate, corneal edema with folds. C, B-scan of the right eye, and D, B-scan of the left eye, revealed shortened ocular axis, bilateral retinochoroidal folds. E, Funduscopy photograph of the right eye, and F, funduscopy photograph of the left eye, indicated retinal detachment, choroidal detachment, and extensive retinochoroidal folds. G, Funduscopy photograph at 2-week of the right eye and, H, funduscopy photograph at 2-week follow-up of the left eye demonstrated optic disk swelling and flattened retinas. I, Fluorescein fundus angiography at 2-week follow up of the right eye, and J, fluorescein fundus angiography at 2-week follow up of the left eye showed hyperfluorescence of the disc and fluorescein leakage.

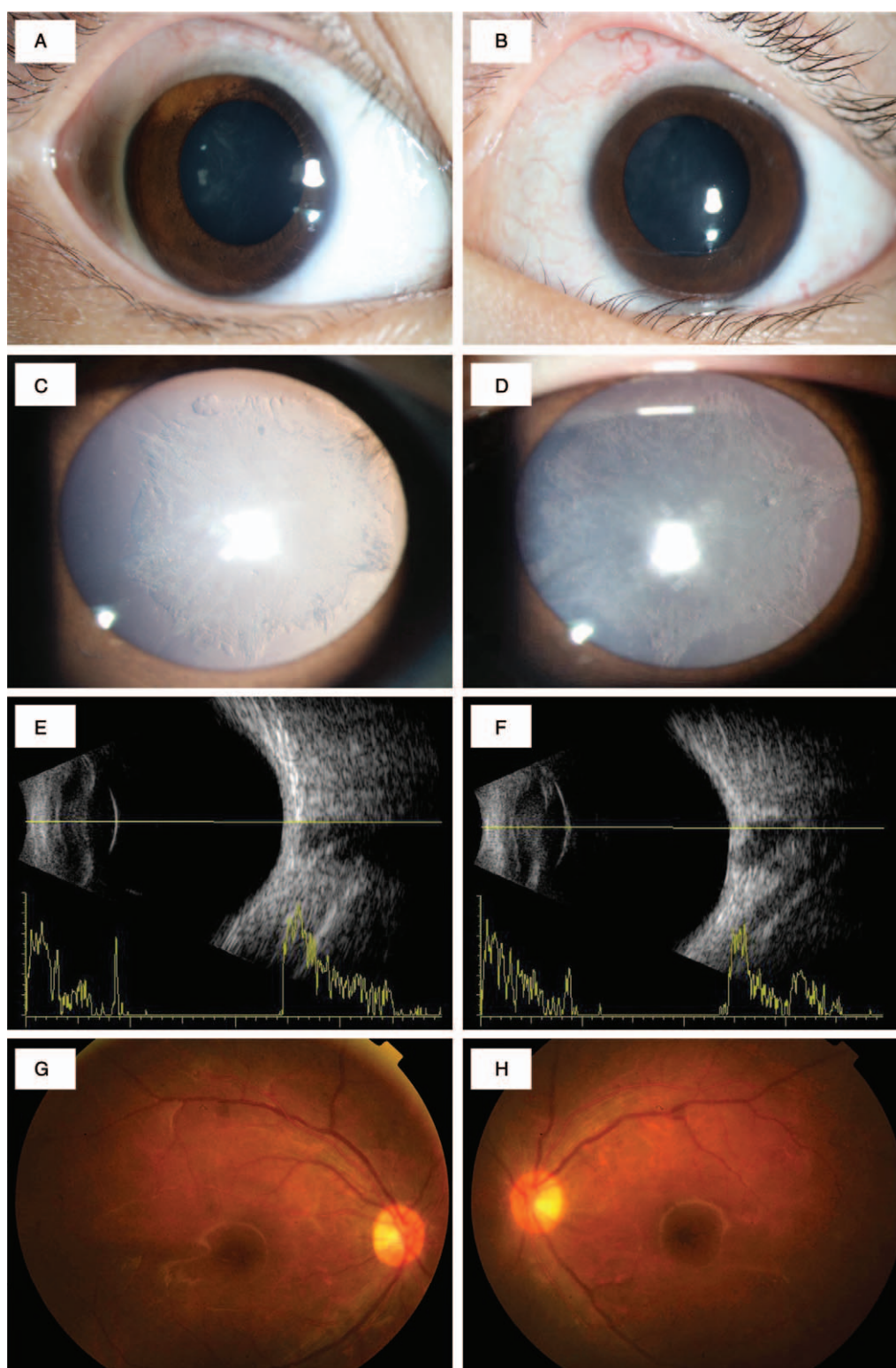


Figure 2. A and B, Photographs at 2-month follow-up suggested inflammation resolved. Both eyes looked normal. C and D, photographs of the anterior segment at 2-month follow-up showed posterior subcapsular opacities. E and F, B-scans at 2-month follow-up revealed normal axial length and normal retinas. G and H, Funduscopic photographs at 2-month follow-up showed normal retinas.

Two days after we started her on intravenous dexamethasone (30 mg/d for 4 days and 15 mg/d for 5 days), the patient was referred to the ophthalmology clinic presenting with ophthalmodynia, photophobia, and enophthalmos. Visual acuity was

HM/5 cm OU. Ocular B-scan showed severe reduction of ocular volume and bilateral chorioretinal folds. Axial length was 15.6 mm OD and 16.1 mm OS (Fig. 1C and D). Intraocular pressure (IOP) was extremely low in OU (unmeasurable). Ophthalmology

examination showed conjunctival edema and injection without exudate, corneal edema with folds (Fig. 1A and B), lens opacity, 3+ aqueous flare, and 1+ keratin precipitates.

Fundus photography suggested retinal detachment, choroidal detachment, and extensive chorioretinal folds (Fig. 1E and D). A fluorescein fundus angiography was obtained 2 weeks later when the ocular inflammation slightly resolved, which revealed hyperfluorescence of the disc and fluorescein leakage (Fig. 1I and J).

Tobramycin dexamethasone eye drops and atropine eye drops were given. The fever and rash started to resolved gradually 4 days after the administration of dexamethasone. She was discharged after 15 days of hospitalization. At follow-ups, retinas gradually flattened and axial length recovered to normal. At 2-month follow-up, visual acuity recovered to 20/50 OU. IOP was normal in OU. Fundus photography revealed normal retinas (Fig. 2G and H). Ophthalmology examination showed posterior subcapsular opacities and normal retinas (Fig. 2C and D). At 24-month follow-up, visual acuity recovered to 20/25 OU (Fig. 2).

3. Discussion

To the best of our knowledge, this is the first documented case of incomplete Kawasaki disease with severe reduction of ocular volume, cataracts, retinal vasculitis, and retinal detachment. We report this case wishing to raise the awareness of posterior segment involvement, especially retinal lesions in Kawasaki disease.

Our patient showed 2 features. She is an older child (10 years old) who didn't present with all the typical symptoms of Kawasaki disease. Despite the correct diagnosis and treatment, her fever was poorly controlled. She had resistance to intravenous immunoglobulin. The devastating enophthalmos, ocular hypotony, and retinal detachment could very likely be the result of the prolonged course of systemic inflammation.

We learned 3 lessons from our practice. The local use of dexamethasone in the eye was effective in our patient. Surgical intervention might not be necessary, given the ocular inflammation could resolve itself as the systemic condition improves. Unnecessary surgeries might cause further damage. Whether the patient needs a surgery depends largely on the evaluation and judgment of the ophthalmologist. Monitoring the eye and the fundus routinely, while treating Kawasaki disease systemically, benefits patients with retinal lesions,^[4] and potentially prevents the ocular inflammation progress to vision loss.

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